Hydrocephalus definition

Hydrocephalus is a common disorder of cerebrospinal fluid (CSF) physiology resulting in abnormal expansion of the cerebral ventricles.

In an attempt to be as inclusive as possible, Raimondi [1] defined cerebrospinal fluid (CSF) as all fluid within the intracranial compartment except blood. With that starting point, hydrocephalus was defined as any increase in CSF within the intracranial compartment, including edema and hydrocephalus ex vacuo. This straightforward definition allowed many pathologies not universally recognized as hydrocephalus by researchers, such as vasogenic edema and cytotoxic edema, to be incorporated into CSF pathophysiology. This attempted definition did not gain widespread acceptance, partially due to its failure to lead to changes in understanding or treatment of the conditions that were included in the definition [2].

In an extensive study supported by the Ministry of Health and Welfare of Japan, Mori and colleagues reviewed a database of 1450 patients with hydrocephalus to develop a classification that would allow outcome to be predicted in individual cases. They excluded tumoral hydrocephalus and defined eight subtypes of hydrocephalus in relationship to the time of onset and severity of brain malformation or injury. They proposed that one of the diagnoses should be “intractable hydrocephalus.” They submitted criteria for this tragic condition for which treatment is futile. The general results of the study were that hydrocephalus is not a disease but a symptom or sign that relates to CSF dynamics [3] [4].

Rekate et al., proposed the following definition of hydrocephalus: “Hydrocephalus is an active distension of the ventricular system of the brain resulting from inadequate passage of CSF from its point of production within the cerebral ventricles to its point of absorption into the systemic circulation.” The elements of this definition are simple and therefore, the number of processes included within it is limited and manageable. Hydrocephalus is an active condition. It is a process that can be demonstrated on neuroimaging studies, and the definition suggests that there is a common underlying cause for its different manifestations: a mismatch between CSF production and its absorption.

Because the definition requires an active process, it excludes brain atrophy or ex vacuo hydrocephalus. Similarly, because this definition requires ventricular distention (ventriculomegaly), it excludes conditions in which there is a failure of CSF absorption such as pseudotumor cerebri (also called benign intracranial hypertension) and normal volume hydrocephalus, this latter being limited to patients shunted during infancy, but found to have increased intracranial pressure (ICP) without ventricular distension at the time of shunt failure [5]. The exclusion of these two conditions will provoke especially energetic debate. It is clear that they are close relatives of hydrocephalus as they are associated with high intracranial pressure and are caused by an increase in the resistance to flow of CSF. They are excluded from the above definition because of the absence of ventricular dilatation. This is a point for general discussion.

The definition does not require an increased ICP even though that condition usually co-exists. Therefore, the conditions of idiopathic and secondary normal pressure hydrocephalus (NPH) are

included. Acceptance of this definition, or one that incorporates these concepts, offers researchers and clinicians a common language and the opportunity to develop a useful classification system, which should be the goal of a future consensus conference.

The proposed definition requires no specific pathologic process. It does not require a researcher to abandon commitment to a bulk flow concept of CSF dynamics or a concept of pulsatility. It also does not require a commitment to an ability to define how CSF is produced, how it is absorbed, or where it is absorbed. It is a point of departure that allows studies on basic aspects of pathophysiology to be incorporated into the definition [6].

1)
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